Complete Summary

GUIDELINE TITLE

Hearing assessment in infants and children: recommendations beyond neonatal screening.

BIBLIOGRAPHIC SOURCE(S)

Cunningham M, Cox EO. Hearing assessment in infants and children: recommendations beyond neonatal screening. Pediatrics 2003 Feb;111(2):436-40. [14 references] PubMed

GUIDELINE STATUS

This is the current release of the guideline.

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COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Hearing loss in infants and small children

GUIDELINE CATEGORY

Diagnosis
Evaluation

Risk Assessment
Screening

CLINICAL SPECIALTY

Family Practice
Nursing
Otolaryngology
Pediatrics

INTENDED USERS

Advanced Practice Nurses
Health Care Providers
Hospitals
Nurses
Physician Assistants
Physicians
Speech-Language Pathologists

GUIDELINE OBJECTIVE(S)

To present recommendations regarding recognition of infants and children at risk for hearing loss, evaluation of infants and children for hearing loss, and proper referral for treatment

TARGET POPULATION

Infants and children

INTERVENTIONS AND PRACTICES CONSIDERED

Risk Assessment and Physical Examination

1. History (in utero and developmental)
2. Questionnaires and checklists (considered, but not recommended)
3. Physical exam (including head and neck exam and eardrum exam)
4. Examination after repeated or chronic otitis media with effusion using pneumatic otoscopy and tympanometry

Audiologic Testing

1. Newborn hearing screening
2. Automated auditory brainstem response (ABR)
3. Evoked otoacoustic emissions (OAE)
4. Behavioral pure tone audiometry
5. Conditioned oriented responses (CORs)
6. Visual reinforced audiometry (VRA)
7. Play audiometry
8. Conventional screening audiometry

Referral to Pediatric Otolaryngologists and Audiologists and Speech and Language Pathologists

MAJOR OUTCOMES CONSIDERED

- Incidence of significant hearing loss in newborns and infants
- Risk of developing hearing loss in infants and children
- Accuracy of audiologic screening tests

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Not stated

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not stated

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Risk Indicators for Congenital or Acquired Hearing Loss

- Most children with congenital hearing loss have hearing impairment at birth and are potentially identifiable by newborn and infant hearing screening. However, some congenital hearing loss may not become evident until later in childhood. Hearing impairment also can be acquired during infancy or childhood for various reasons. All older infants and children should be screened for risk factors involving hearing problems. Every child (birth through 24 months of age) found to have 1 or more of the following high-risk indicators should be followed and periodically screened for late-onset congenital or acquired hearing loss.

  Birth to 28 days

  - Family history of sensorineural hearing loss (SNHL), presumably congenital
  - In utero infection associated with SNHL (e.g., toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis)
  - Ear and other craniofacial anomalies including abnormalities of the eardrum, heterochromia of the irises, malformation of the auricle or ear canal, dimpling or skin tags around the auricle, cleft lip or palate, asymmetry or hypoplasia of the facial structures, and microcephaly
  - Hyperbilirubinemia at levels requiring exchange transfusion
  - Birth weight less than 1500 g
  - Bacterial meningitis
  - Low Apgar scores: 0-3 at 5 min; 0-6 at 10 min
  - Respiratory distress (e.g.,, meconium aspiration)
  - Prolonged mechanical ventilation for more than 10 d
  - Ototoxic medication (e.g., gentamicin) administered for more than 5 d or used in combination with loop diuretics
  - Physical features or other stigmata associated with a syndrome known to include SNHL (e.g., Down syndrome, Waardenburg syndrome)

  29 days to 24 months

    - Parental or caregiver concern about hearing, speech or language, and/or developmental delay
    - Any of the newborn risk factors listed above
    - Recurrent or persistent otitis media with effusion (OME) for at least 3 mo
    - Head trauma with fracture of temporal bone
    - Childhood infectious diseases associated with SNHL (e.g., meningitis, mumps, measles)
    - Neurodegenerative disorders (e.g., Hunter syndrome) or demyelinating diseases (e.g., Friedreich ataxia, Charcot-Marie-Tooth syndrome)
- Although questionnaires and checklists are useful in identifying a child at risk for hearing loss, studies have shown that only 50% of children with hearing loss are identified by the comprehensive use of such questionnaires. Therefore, periodic objective assessment of the hearing of all children should be performed.
- If a parent or caregiver is concerned that a child cannot hear, the pediatrician needs to assume that such is true until the child has been evaluated objectively. Any parental concern should be taken seriously, and formal hearing evaluation should be performed.
- Failure to achieve the following milestones by expected age ranges might relate to hearing loss that necessitates audiologic testing:

  <u>Birth to 3 months</u>

    - Startles to loud noise
    - Awakens to sounds
    - Blinks or widens eyes in response (reflex) to noises

  <u>3-4 months</u>

    - Quiets to mother's voice
    - Stops playing, listens to new sounds
    - Looks for source of new sounds not in sight

  <u>6-9 months</u>

    - Enjoys musical toys
    - Coos and gurgles with inflection
    - Says "mama"

  <u>12-15 months</u>

    - Responds to his or her name and "no"
    - Follows simple requests
    - Uses expressive vocabulary of 3 to 5 words
    - Imitates some sounds

  <u>18-24 months</u>

    - Knows body parts

- Uses expressive vocabulary 2-word phrases (minimum of 20 to 50 words)
- 50% of speech intelligible to strangers

By 36 months

- Uses expressive vocabulary of 4- to 5-word sentences (approximately 500 words)
- Speech is 80% intelligible to strangers
- Understands some verbs

Physical Examination

A thorough physical examination is an essential part of evaluating a child for hearing loss. Findings on a head and neck examination associated with hearing impairment include heterochromia of the irises, malformation of the auricle or ear canal, dimpling or skin tags around the auricle, cleft lip or palate, asymmetry or hypoplasia of the facial structures, and microcephaly. Hypertelorism and abnormal pigmentation of the skin, hair, or eyes also may be associated with hearing loss, as in Waardenburg syndrome. Abnormalities of the eardrum should alert the physician to the possibility of hearing impairment. A leading cause of acquired hearing impairment is otitis media with effusion (OME). Temporary hearing loss has been demonstrated during episodes of acute otitis media. The child with repeated or chronic OME is at high risk of acquired hearing impairment and should undergo hearing evaluation. Pediatricians should be familiar with pneumatic otoscopy and tympanometry as useful diagnostic tools in the management of OME.

Tools for Objective Hearing Screening

- In addition to universal newborn hearing screening, objective screenings for hearing impairment should be performed periodically on all infants and children in accordance with the schedule outlined in the AAP statement, "Recommendations for Preventive Pediatric Health Care."
- The technology used for hearing screening should be age appropriate. The child also should be comfortable with the testing situation; young children may need preparation.
- Screenings should be conducted in a quiet area where visual and auditory distractions are minimal.
- Screening tests:

  Auditory brainstem response (ABR)

  The automated auditory brainstem response (ABR) instrument measures ABRs at frequencies greater than 1000 Hz with a broadband click stimulus in each ear. The testing instrument incorporates a built-in artifact rejection for myogenic, electrical, and environmental noise interference, which ensures that data collection is halted if testing conditions are unfavorable. The automated screener provides a pass-fail report; no test interpretation by an audiologist is required. Automated ABR can test each ear individually and can be performed on children of any age. Motion artifact interferes with test results. For this reason, the test is performed best in infants and young

children while they are sleeping or, if necessary, sedated. The ABR is currently used in many newborn programs.

Evoked otoacoustic emissions (OAE)

Evoked otoacoustic emissions (OAE) are acoustic signals generated from within the cochlea that travel in a reverse direction through the middle ear space and tympanic membrane out to the ear canal. These signals are generated in response to clicks or tone bursts. The signals may be detected with a sensitive microphone/probe system placed in the external ear canal. The OAE test allows for individual ear assessment, is performed quickly at any age, and is not dependent on whether the child is asleep or awake. Motion artifact does interfere with test results. The OAE is an effective screening measure for inner and middle ear abnormalities, because at hearing thresholds of 30 dB or higher, there is no OAE response. The OAE test does not further quantify hearing loss or hearing threshold level. The OAE also does not assess the integrity of the neural transmission of sound from the eighth nerve to the brainstem and, therefore, will miss auditory neuropathy and other neuronal abnormalities. Infants with such abnormalities will have normal OAE test results but abnormal ABR test results.

Limitations of ABR and OAE tests

The ABR and OAE are tests of auditory pathway structural integrity but are not true tests of hearing. Even if ABR or OAE test results are normal, hearing cannot be definitively considered normal until a child is mature enough for a reliable behavioral audiogram to be obtained. Behavioral pure tone audiometry remains the standard for hearing evaluation. Hearing thresholds at specific frequencies can be determined and the degree of hearing impairment can be assigned. If there are distractions or the room is not soundproof, pure tone audiometry in the office should be considered solely a screening test.

Conditioned oriented responses (CORs) or visual reinforced audiometry (VRA)

Children as young as 9 to 12 months can be screened by means of conditioned oriented responses (CORs) or visual reinforced audiometry (VRA). Both of these techniques condition the child to associate speech or frequency-specific sound with a reinforcement stimulus, such as a lighted toy or dancing animal. The VRA is a more sophisticated and accurate form of COR requiring a soundproof room and is typically performed by an audiologist.

Play audiometry

Children 2 to 4 years of age are tested more appropriately by play audiometry. These children are conditioned to respond to an auditory stimulus through play activities, such as dropping a block when a sound is heard through earphones.

Conventional screening audiometry

For children 4 years and older, conventional screening audiometry can be used. The child is asked to raise the right or left hand when a sound is heard in the respective ear. The test should be performed in a quiet environment using earphones, because ambient noise can affect test performance significantly, especially at lower frequencies (i.e., 500 and 1000 Hz). Each ear should be tested at 500, 1000, 2000, and 4000 Hz. Air conduction hearing threshold levels of >20 dB at any of these frequencies indicate possible impairment.

Audiometric evidence of hearing loss should be substantiated by repeat screening. Earphones should be removed and repositioned, and instructions should be carefully repeated to the child to ensure proper understanding and attention to the test. A child whose repeat test shows hearing thresholds >20 dB at any of these frequencies, especially if there is no pathologic abnormality of the middle ear on physical examination, should be referred for formal hearing testing. Children with unilateral or mild hearing loss also should be further evaluated; studies show such children to be similarly at risk for adverse communication skills as well as difficulties with social, emotional, and educational development.

- The results of hearing screening and ear examinations should be explained carefully to parents. The child's chart should be marked clearly to facilitate tracking of appropriate referrals, developmental skills, and school performance.

Hearing Referral Resources

- Pediatricians should be familiar with the referral resources available in their community for hearing-impaired children.
- Pediatric otolaryngologists and audiologists and speech and language pathologists with special training and experience in treating children should be consulted for specific diagnosis, counseling, and treatment. The primary care pediatrician and the otolaryngologist should collaborate to refer the child for comprehensive educational counseling and treatment services. Communication among professionals caring for a hearing-impaired child is essential to ensure appropriate case management.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

Supportive evidence is outlined in the Joint Committee on Infant Hearing´s "Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs," which was endorsed by the guideline developer.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Prevention of adverse consequences of hearing loss in infancy and childhood (lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, and emotional difficulties) and facilitation of language acquisition in children with hearing loss.

Subgroups Most Likely to Benefit:

Patients identified with hearing loss in the first 6 months of life

POTENTIAL HARMS

Not stated

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

The recommendations in this statement do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Staying Healthy

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2003 Feb

GUIDELINE DEVELOPER(S)

American Academy of Pediatrics - Medical Specialty Society

SOURCE(S) OF FUNDING

American Academy of Pediatrics

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Committee on Practice and Ambulatory Medicine

Section on Otolaryngology and Bronchoesophagology

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

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GUIDELINE AVAILABILITY

Electronic copies: Available from the American Academy of Pediatrics (AAP) Policy Web site.

Print copies: Available from American Academy of Pediatrics, 141 Northwest Point Blvd., P.O. Box 927, Elk Grove Village, IL 60009-0927.

AVAILABILITY OF COMPANION DOCUMENTS

None available

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on May 15, 2003. The information was verified by the guideline developer on June 9, 2003.

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